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Clinical vignette

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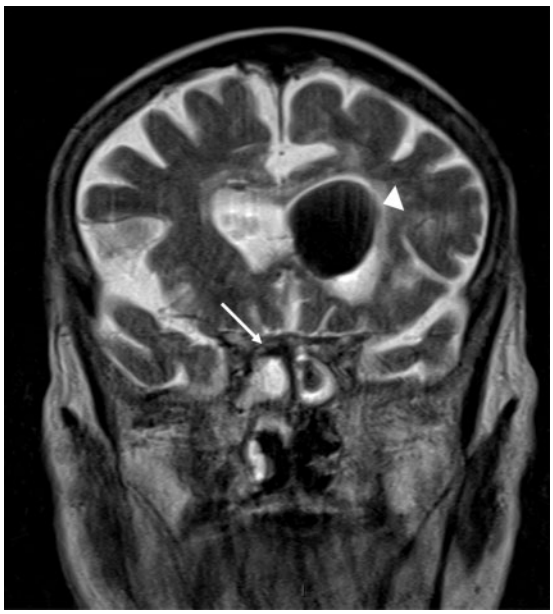
Pneumocephalus due to granulomatosis with polyangiitis

A 67-year-old woman affected by granulomatosis with polyangiitis (GPA) presented with a flare of lung disease and ulceration of the lower limb without active sinusitis. She had a 9-year history of GPA with pulmonary and ENT involvement, the latter leading to left eye enucleation (histological findings suggestive of GPA). She had initially been treated with CYC (6 months) and was currently on steroids and MTX. Laboratory studies revealed raised inflammatory markers and cANCA (PR3) positivity. Thoracic CT showed progression of bilateral ground-glass opacities; bronchoalveolar lavage excluded infections. Methylprednisolone 1 mg/kg was introduced with clinical improvement. However, she gradually developed episodic headache and a mild, fluctuant altered mental status with confusion and delayed verbal response without other neurological features. A coronal T2-weighted MRI (Fig. 1) revealed a previously known, unmodified ischaemic lesion and the presence of extensive pneumocephalus secondary to cerebrospinal fluid leakage through multiple discontinuities in the ethmoid bone due to chronic localization of GPA to the anterior skull base (without other risk factors for development of pneumocephalus). Pneumocephalus due to cerebrospinal fluid fistula is usually secondary to trauma or surgery [1], but to our knowledge has never been reported as a complication of skull base involvement in GPA.

FIG. 1 Coronal T2-weighted MRI revealing the presence of cerebrospinal fluid in the nasal and paranasal cavities (arrow) and air in the left frontal horn of the ventricle (arrowhead)



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